

Postsplenectomy sepsis 10 years or more after operation

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SUMMARY Three new cases (two fatal) of postsplenectomy sepsis occurring 14, 25, and 26 years after operation for hereditary spherocytosis are described. There are now 25 cases in the published work in which this complication occurred 10 or more years after operation, 14 of which were fatal. The mean age of onset is 37 years. The features of the disease are similar to those seen in other postsplenectomy infections, and pneumococcal infection was responsible in 19 cases (76%). The major predisposing illnesses were trauma, hereditary spherocytosis, and idiopathic thrombocytopenia.

The risk of severe, sometimes fatal, infection after splenectomy is well recognised.¹⁻⁷ It seems to be more common in children, but occurs at all ages. The risk is greatest in the early months and years after operation.² It may never disappear entirely, however, and one case occurring 45 years after operation has been described.⁸ Several isolated case reports and small series have recorded late infections after splenectomy (Table 1), but this paper summarises the previous cases of postsplenectomy infection 10 or more years after operation. Three new cases are described, in which infection developed 14, 25, and 26 years after splenectomy; two of the patients died. The total number of reported cases of postsplenectomy infection 10 or more years after operation is now 25.

Case reports

CASE 1

An 8 year old boy had his spleen removed for hereditary spherocytosis in 1951. Twenty six years later, aged 33, he was admitted to hospital with fever and malaise of sudden onset. He rapidly went cold and blue, lost consciousness, and died. A coroner's necropsy showed adrenal haemorrhage due to meningococcal septicaemia.

CASE 2

A 16 year old girl had an elective splenectomy for hereditary spherocytosis in 1964. Fourteen years later, aged 30, she developed a sudden illness with

headache, purpura, and collapse. She had thrombocytopenia and afibrinogenaemia. The cerebrospinal fluid was heavily blood stained and grew pneumococci. She died the following day. The diagnosis was a consumption coagulopathy due to pneumococcal meningitis. No necropsy was performed.

CASE 3

A 4 year old girl underwent an elective splenectomy for hereditary spherocytosis. Twenty five years later she was admitted to hospital with pneumococcal meningitis. There were no complications and she made a full recovery.

Discussion

These three case histories summarise the features of post-splenectomy sepsis^{1,2}: there is a high rate of pneumococcal infection, and a lower rate of meningococcal disease; disseminated intravascular coagulation and adrenal haemorrhage are prominent complications; and fatal infection develops with dramatic suddenness.

All three patients reported here had hereditary spherocytosis; this is not coincidental. All were parents of children who attend haematology clinics at children's hospitals in Manchester. There are about 50 such families registered, and the children may attend with parents, grandparents, and other relatives. As the disease is inherited as an autosomal dominant disease, a large number of relatives have been seen, many of whom have had splenectomies.

Including these three cases, there are now 25 pub-

Table 1 Patients with sepsis 10 or more years after splenectomy

Sex	Age	Diagnosis	Organisms	Years after operation	Presentation	Outcome	Reference no
F	17	Spherocytosis	Pneumococcus	14	DIC	Died	9
F	34	ITP	Pneumococcus	15	Endocarditis	Survived	10
M	18	ITP	Pneumococcus	10	DIC	Died	10
F	32	Trauma	Pneumococcus	12	DIC	Died	11
F	34	ITP	Pneumococcus	15	DIC	Died	12
M	36	Trauma	<i>H influenzae</i>	15	Meningitis	Survived	13
F	44	Gastrectomy	Pneumococcus	10	DIC	Died	14
M	47	Trauma	Pneumococcus	25	DIC	Died	15
M	23	Trauma	Pneumococcus	10	DIC	Died	16
F	43	Anaemia, cause unknown	Pneumococcus	21	Septicaemia	Died	17
M	27	Trauma	Pneumococcus	10	Adrenal haemorrhage	Died	18
F	47	Spherocytosis	Meningococcus	10	Meningitis	Survived	19
M	58	ITP	Pneumococcus	23	Septicaemia	Survived	20
M	51	Trauma	Pneumococcus	45	Septicaemia	Died	8
M	28	Trauma	Unidentified*	28	Septicaemia	Survived	21
M	33	Trauma	Pneumococcus	13	DIC	Died	22
F	46	Trauma	Pneumococcus	31	Septicaemia	Survived	22
F	49	SLE and thrombocytopenia	Pneumococcus	25	Septicaemia	Died	23
M	40	ITP	Salmonella	11	Septicaemia	Survived	23
M	42	Trauma	Streptococcus	19	Septicaemia	Survived	23
F	31	Mononucleosis	Pneumococcus	11	Septicaemia	Survived	23
M	44	Trauma	Pneumococcus	20	Meningitis	Survived	24
M	33	Spherocytosis	Meningococcus	26	Septicaemia W-F syndrome	Died	Present series
F	30	Spherocytosis	Pneumococcus	14	Meningitis	Died	Present series
F	29	Spherocytosis	Pneumococcus	25	Meningitis	Survived	Present series

ITP = idiopathic thrombocytopenic purpura.

SLC = systemic lupus erythematosus.

DIC = disseminated intravascular coagulation.

WF syndrome = Waterhouse-Friderichsen syndrome.

*The organism was repeatedly isolated but proved unidentifiable.

lished cases of postsplenectomy sepsis developing 10 or more years after the operation. Thirteen were men and 12 were women; 14 died and 11 survived. In 18 cases (72%) pneumococcus was implicated. Reasons for operation include trauma (11 cases), idiopathic thrombocytopenic purpura (5 cases), and hereditary spherocytosis (5 cases) (Table 1).^{8,9-24}

The deaths occurred suddenly in patients in early middle age; the mean age at infection was 37 years. None had been given any prophylactic treatment; indeed, the risk of infection in adults was not recognised until the late sixties, and in most cases the operation had been performed earlier. The risk is still not widely known, and what steps we should take to reduce the risk remain a matter for discussion. Furthermore, none of the adult patients seen with their children in the paediatric haematology clinics had ever been offered any prophylactic treatment, and none was aware of the risk of infection.

There are wide variations in the time at which patients are at risk (Table 2).^{23,25-30} There is no doubt that the risk is maximal in the early years after operation, but the present data indicate that, for some patients, the risk never disappears entirely. However, only patients treated for trauma,

haemolytic anaemia, and idiopathic thrombocytopenic purpura appear to be at risk. An analysis of all cases undergoing splenectomy in the North Western Regional Health Authority's hospitals between 1975 and 1982 showed that these diagnoses accounted for only 29% of splenectomies in a total of 2066 operations. Although malignant disease, cirrhosis, incidental splenectomy at gastrectomy, and other disorders account for most splenectomies, the poor survival rates from most of these disorders account for their absence in this list of long term post splenectomy sepsis.

Antipneumococcal and other vaccines^{31,32} or penicillin³ are being evaluated for prophylaxis. Their

Table 2 Time to sepsis after splenectomy

	Reference number
Majority within six years	Bisno and Freeman 1970 25
80% within four years	Eraklis and Filler 1972 26
75% within two years	Likhite 1976 27
No difference	Ramsay and Bouskill 1976 28
Majority within three years	Walker 1976 29
91% within six weeks	Ein <i>et al</i> 1977 30
Half within three years	Askergren and Björkholm 1980 23

value has not yet been established, and cases have been reported of fatal infections in patients with sickle cell disease (who are similarly predisposed to sepsis) after pneumococcal vaccine^{33,34} and in a splenectomised child given both pneumococcal vaccine and penicillin.³⁵ It has not yet been established how often, if at all, pneumococcal vaccine needs to be given. At present the manufacturers recommend not less than five years between revaccination; earlier repeats risk unpleasant reactions. There is, however, a well established precedent for the lifelong use of penicillin for prophylaxis by patients at risk of rheumatic heart disease.³⁶ Lifelong penicillin prophylaxis could be extended to splenectomy patients. The present data suggest that long term prophylaxis is needed only for patients whose splenectomy is performed for trauma, congenital haemolytic anaemia, and idiopathic thrombocytopenic purpura. It does not seem likely, however, that patients would continue to take penicillin for many years or that physicians would prescribe it. Nor is it clear which doctor should be responsible. My investigations show that the risk persists and extend Ramsay's finding³⁷ that in adults many cases of postsplenectomy sepsis occur more than five years after operation. Two or three years' postoperative penicillin is unlikely to be effective. Any pyrexia of undetermined origin in a splenectomised patient demands blood culture and treatment with ampicillin.³⁷ Hospital junior staff, who have first contact with emergencies, must therefore be made aware of the problem. At the same time, we should advise our patients to wear a locket giving case details and guidance to treatment and give them a medical card indicating the diagnosis and clearly stating the risk of postsplenectomy sepsis and its management.

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